Journal of Neurology & Experimental Neuroscience

https://doi.org/10.17756/jnen.2019-060

Case Report

Open Access

Nonconvulsive Status Epilepticus (NCSE) Presenting as Acute Confusional State

Suellen Li[1*], Andrea Loggini[2] and John H Pula[3]

[1]*Department of Medicine, Massachusetts General Hospital, Boston, MA, USA*
[2]*Department of Neurology, University of Chicago, Chicago, IL, USA*
[3]*Department of Neurology, NorthShore University Health System, Evanston, IL, USA*

[*]Correspondence to:
Dr. Suellen Li, MD
Massachusetts General Hospital Department of Medicine, Boston, MA, USA
E-mail: suellenli62@gmail.com

Received: August 29, 2019
Accepted: November 30, 2019
Published: December 02, 2019

Citation: Li S, Loggini A, Pula JH. 2019. Nonconvulsive Status Epilepticus (NCSE) Presenting as Acute Confusional State. *J Neurol Exp Neurosci* 5(2): 100-102.

Published by United Scientific Group

Abstract

Background: Nonconvulsive status epilepticus (NCSE) is a critical neurological condition that is commonly under-recognized due to its myriad variability of clinical presentation. High-risk groups include epileptic patients, the elderly, and critically ill populations. Diagnosis is supported with continuous EEG monitoring. Its treatment can follow a protocol created specifically for status epilepticus, with intravenous benzodiazepines usually given as first line therapy.

Clinical Case: This case is of a 42-year-old female with distant history of unspecified epilepsy with absence spells off anti-epileptic medications for a prolonged time, brought to the emergency room after being found acutely confused, wandering at a train station. On initial neurological evaluation, she was disoriented, but alert. Initial workup, including brain MRI with gadolinium, electrolyte, toxicity screen, and evaluation for infection was normal. EEG was initiated and immediately demonstrated generalized continuous polyspike-wave discharges, compatible with non-convulsive status epilepticus. Intravenous benzodiazepines (lorazepam 2 mg) followed by a loading dose of intravenous fosphenytoin were promptly administered with complete resolution of confusion and slow return to an electrographically normal background rhythm of 9-10 Hz. She was observed for 24 hours and discharged the following day on antiepileptic medication.

Conclusions: Acute confusional state, even in absence of focal neurological findings, must raise the question of NCSE. EEG is a rapid diagnostic tool that should be performed in all suspected cases. Early diagnosis and prompt treatment prevent further clinical deterioration, refractory disease, and irreversible neurological damages. Ready access to EEG testing and interpretation is critical for the timely identification and treatment of these patients.

Introduction

Nonconvulsive status epilepticus (NCSE) is a relatively rare neurological condition defined as a dysfunction in mental status or behavior associated with continuous epileptiform discharges on EEG lasting over 30 minutes [1, 2]. The unified criteria published in the literature include frequency cutoffs for epileptiform discharges, the presence of clinical improvement after administration of an IV antiepileptic medication, ictal phenomena noted during epileptiform activity, spatiotemporal evolution, and a comparison to baseline for patients with known epilepsy. NCSE is further classified into patients with and without coma/stupor [3]. NCSE may be present in a wide array of clinical situations, ranging from coma to patients who are awake but behaving unusually [4].

Some patients with nonconvulsive status epilepticus have a history of epilepsy, often since childhood. Common triggers include stress, lack of sleep, drugs, alcohol, illness, or photophobic stimulation. However, it is less common for NCSE to be the first presentation of seizure [5]. Patients who first present in NCSE have a worse outcome than those with pre-existing epilepsy because de novo NCSE often arises from progressive CNS disorders [1].

NCSE must be differentiated from other mimics including stroke, metabolic and toxic encephalopathy, infections, traumatic encephalopathy, and psychological conditions such as dissociative disorders. Certain clinical features have been shown to be more likely in patients with NCSE, including remote risk factors for seizures, severely altered mental status, and ocular movement abnormalities [6]. The principal differentiation is based on finding epileptiform discharges on EEG. Therefore, EEG should be obtained in all cases in which there is a suspicion for NCSE. The treatment of NCSE can follow a protocol created specifically for status epilepticus, with intravenous benzodiazepines usually given as first line therapy [7].

Case Report

This is the case of a 42-year-old college-educated female with a history of remote unspecified epilepsy with absence spells in childhood who had self-discontinued medications in adolescence (she was unable to recall which medication she had taken). She was brought to the ED by EMS after being found at a train station with altered mental status. She had been traveling out of town and had been scheduled to fly back home the afternoon of admission. When she failed to show up for her flight, police were called and tracked her phone signal to a train station. She was unable to answer how she got there and thought she was at home. She reported multiple times that she had fallen but was unable to provide further details.

Upon arrival to the hospital, she denied pain, loss of consciousness, other symptoms, or recent illness. She was able to identify that she was in the hospital, but thought she was in her home state, and did not know why she was at the hospital.

Per her husband, she had been in her usual health recently and had not had any recent illnesses. She had not been taking medications at home and had not had a seizure since age 12. She had no known history of abuse. She was alert and oriented to self but not to year or location. She did not remember the events of the previous day but remembered being at a baseball game two days prior to presentation. She was last seen normal by a friend on the morning of presentation at breakfast. Further evaluation, including a chest x-ray, CT of the head and cervical spine, MRI of the brain, complete blood count, metabolic panel, cardiac markers, and urine toxicology test was negative. On initial neurological exam, she reported sensing touch on her left side when her right side was being touched.

Neurology was consulted the morning after presentation. Her neurologic exam at that time showed impaired attention, with inability to complete serial 7s or recite the months of the year backwards. She had mildly impaired delayed recall, remembering 2 out of 3 words after 3 minutes. Otherwise, she was answering questions and following commands appropriately. Language comprehension, fluency, rhythm, intonation, repetition, and content were normal. Her fund of knowledge was consistent with her education and her cranial nerves were all intact. Motor exams, including coordination and gait, and sensory exams were unremarkable. She was able to correctly identify the side of her that was being touched.

A 30-minute EEG was immediately requested that revealed Nonconvulsive status epilepticus (Figure 1). The EEG polyspike and wave pattern was characteristic of juvenile myoclonic epilepsy. She was given 2 mg of IV lorazepam and a loading dose of fosphenytoin (20 mg/kg), which resolved the seizure. Her mental status subsequently returned to baseline, which was confirmed by her husband, and she was maintained on fosphenytoin 300 mg three times a day. Post-treatment EEG demonstrated normal background rhythm, and it was continued for 24-hour without reemergence of epileptiform discharges (Figure 2). The trigger was thought to be sleep deprivation and she was counseled on sleep hygiene and medications that may lower the seizure threshold. On discharge, she was prescribed phenytoin 300 mg at night time with outpatient neurology follow up.

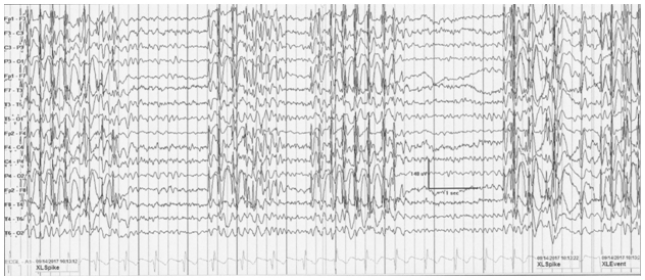

Figure 1: Initial EEG. 17 Channel XLTEK digital EEG demonstrating generalized continuous 3-4 Hz polyspike-wave discharges characteristic of status epilepticus with findings suggestive of juvenile myoclonic epilepsy.

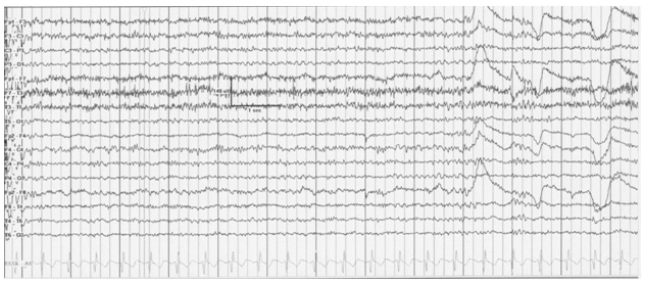

Figure 2: Post-Treatment EEG. 17 Channel XLTEK digital EEG demonstrating normal background rhythm with well-formed, symmetric, and synchronous 9-10 Hz rhythms.

Discussion

NCSE can present in various circumstances and clinical manifestations and the differential for each presentation is broad. Acute confusional state, even in absence of focal neurological findings, must raise the question of NCSE and workup for it has to be promptly initiated when alternative etiologies have been excluded.

EEG is a rapid and inexpensive diagnostic tool that should be performed in all suspected cases. However, it is important to recognize several limitations in the utilization of this ancillary test. Firstly, its utilization is limited by the availability of EEG machines and technicians, especially during nights and weekends. Secondly, the interpretation of EEG is operator dependent and the detection of more subtle findings might be missed in undertrained physicians. Finally, in our current healthcare system, where resource utilization represents an important issue, guidance from a neurology trained physician should be sought in order to avoid ordering unnecessary tests. In addition, training ED providers to recognize the key signs of NCSE may be a strategy that would help more rapidly identify and treat these patients.

In this case, there are several interesting factors, including that this patient with childhood epilepsy and possible findings of juvenile myoclonic epilepsy (JME) on EEG had no reported seizures for several decades while off antiepileptic medications. 3-8% of childhood absence epilepsies evolve into JME, which is characterized by myoclonic jerks [8]. Furthermore, it is unusual that she had no myoclonic activity on exam nor did she have any reported history of myoclonic jerks noted by the patient herself or her family. It is possible that this patient may have had brief seizures that were mild in nature and went unnoticed. Alternatively, it is possible that she truly had no seizures. In fact, 9% of JME patients in a long-term observational study remained seizure free for years off antiepileptic medications [9]. This patient may have been seizure-free for years and had an episode of NCSE triggered by sleep deprivation. NCSE is rare in adult patients with JME and little is known about its frequency outside of case reports, which suggest a frequency of 5.8% with a female predominance [10].

Phenytoin was administered as second line agent to abort refractory NCSE. The administration of it promptly stopped the epileptic activity. The utilization of a sodium channel blocker in an adult with remote history of epilepsy who had been seizure free for many years is controversial, and few other agents might have been considered such as valproic acid or levetiracetam. We decided not to start valproic acid in a woman of childbearing age, nor levetiracetam in a patient who presented with a confusional episode, as further spells might have been confused for levetiracetam toxicity. We also did not have the luxury to initiate a better long-term medication, such as lamotrigine, because she was coming from out of state and we couldn't monitor the stepwise increase in the drug. Thus, the decision was to keep her on phenytoin until care was established in an outpatient environment.

Furthermore, it is important to examine the timing of treatment. Timely treatment of NCSE, while not as critical compared to convulsive status epilepticus, is still related to subsequent prognosis. In one series of 38 NCSE cases, the median time from the initiation of NCSE to control was 16.5 hours [11]. For this patient, over 24 hours had elapsed between when she was last known well and when she achieved control of her NCSE. While 11 of these hours were due to a delay in presentation to the hospital, it took 13 hours from her arrival in the ED to control of her NCSE. This was likely related to the atypical presentation that did not immediately raise concern for ongoing seizure activity, but also due in part to the unavailability of EEG technicians overnight, which resulted in a significant delay in diagnosis and treatment.

For patients with NCSE, triggers such as poor sleep, infection, or toxic exposure should be identified and addressed. Early diagnosis with EEG and prompt treatment prevent further clinical deterioration, refractory disease, and irreversible neurological damages. EEG interpretation should be available in a timely manner to allow for the rapid identification and treatment of NCSE patients. This could be done through early involvement of neurology trained physicians in the care of these patients.

References

1. Sutter R, Rüegg S, Kaplan PW. 2012. Epidemiology, diagnosis, and management of nonconvulsive status epilepticus. *Neuro Clin Pract* 2(4): 275-286. https://doi.org/10.1212/CPJ.0b013e318278be75
2. Trinka E, Cock H, Hesdorffer D, Rossetti AO, Scheffer IE, et al. 2015. A definition and classification of status epilepticus-Report of the ILAE Task Force on Classification of Status Epilepticus. *Epilepsia* 56(10): 1515-1523. https://doi.org/10.1111/epi.13121
3. Beniczky S, Hirsch LJ, Kaplan PW, Pressler R, Bauer G, et al. 2013. Unified EEG terminology and criteria for nonconvulsive status epilepticus. *Epilepsia* 54 Suppl 6: 28-29. https://doi.org/10.1111/epi.12270
4. Holtkamp M, Meierkord H. 2011. Nonconvulsive status epilepticus: a diagnostic and therapeutic challenge in the intensive care setting. *Ther Adv Neurol Disord* 4(3): 169-181. https://doi.org/10.1177/1756285611403826
5. Tomson T, Lindbom U, Nilsson BY. 1992. Nonconvulsive status epilepticus in adults: thirty-two consecutive patients from a general hospital population. *Epilepsia* 33(5): 829-835. https://doi.org/10.1111/j.1528-1157.1992.tb02190.x
6. Husain AM, Horn GJ, Jacobson MP. 2003. Non-convulsive status epilepticus: usefulness of clinical features in selecting patients for urgent EEG. *J Neurol Neurosurg Psychiatry* 74(2): 189-191. https://doi.org/10.1136/jnnp.74.2.189
7. Manno EM. 2011. Status epilepticus: current treatment strategies. *Neurohospitalist* 1(1): 23-31. https://doi.org/10.1177/1941875210383176
8. Renganathan R, Delanty N. 2003. Juvenile myoclonic epilepsy: under-appreciated and under-diagnosed. *Postgrad Med J* 79(928): 78-80. https://doi.org/10.1136/pmj.79.928.78
9. Höfler J, Unterberger I, Dobesberger J, Kuchukhidze G, Walser G, et al. 2014. Seizure outcome in 175 patients with juvenile myoclonic epilepsy--a long-term observational study. *Epilepsy Res* 108(10): 1817-1824. https://doi.org/10.1016/j.eplepsyres.2014.09.008
10. Dziewas R, Kellinghaus C, Lüdemann P. 2002. Nonconvulsion status epilepticus in patients with juvenile myoclonic epilepsy: types and frequencies. *Seizure* 11(5): 335-339. https://doi.org/10.1053/seiz.2001.0611
11. Gutiérrez-Viedma Á, Parejo-Carbonell B, Cuadrado ML, Serrano-García I, Abarrategui B, et al. 2018. The relevance of timing in nonconvulsive status epilepticus: a series of 38 cases. *Epilepsy Behav* 82: 11-16. https://doi.org/10.1016/j.yebeh.2018.02.029